Health Care Provider Fact Sheet

Disease Name Hemoglobin C Disease

Alternate Name(s) Sickle Cell Hemoglobin C Disease

Acronym Hb S/C

Disease Classification Hemoglobinopathy

Symptom onset May be asymptomatic.

Symptoms Any sign of illness in an infant with sickling disease is a potential medical

emergency. Acute and chronic tissue injury can occur when sickled cells cause vascular occlusion. Sickling diseases can cause severe pain anywhere in the body, but most often in the hands, arms, chest, legs and feet. Complications may include, but are not limited to, the following: sepsis, acute chest syndrome, hand-and-foot syndrome, splenic

sequestration crisis, aplastic crisis, stroke and painful episodes.

Natural history without treatment Infants with hemoglobin C disease are vulnerable to serious bacterial

infections that can be life threatening.

Natural history with treatmentReduced mortality and morbidity with penicillin prophylaxis.

Treatment The National Institutes of Health clinical guidelines for management of

sickle cell disease state, "Penicillin prophylaxis should begin by 2 months of age for infants with suspected sickle cell anemia, whether or not the definitive diagnosis has been established." Antibiotic therapy should

continue until at least 5 years of age.

Emergency Medical Treatment See sheet from American College of Medical Genetics (attached) or for more

information, go to website:

http://www.acmg.net/StaticContent/ACT/ACT-sheet_HBSC_FSC.pdf

Inheritance Autosomal recessive

General population incidence Affects 2 to 3% of African American in the United States.

OMIM Link http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=603903

Genetests Link www.geneclinics.org

Support Group Sickle Cell Information Center

http://www.scinfo.org/

Sickle Cell Disease Association of America, Inc.

http://www.sicklecelldisease.org

4-26-2010 Update

